Genito-urinary Malformations consequent on Pelvic Deformities.

By John Yule Mackay, M.D., Senior Demonstrator of Anatomy, University of Glasgow. (Plate VII.)

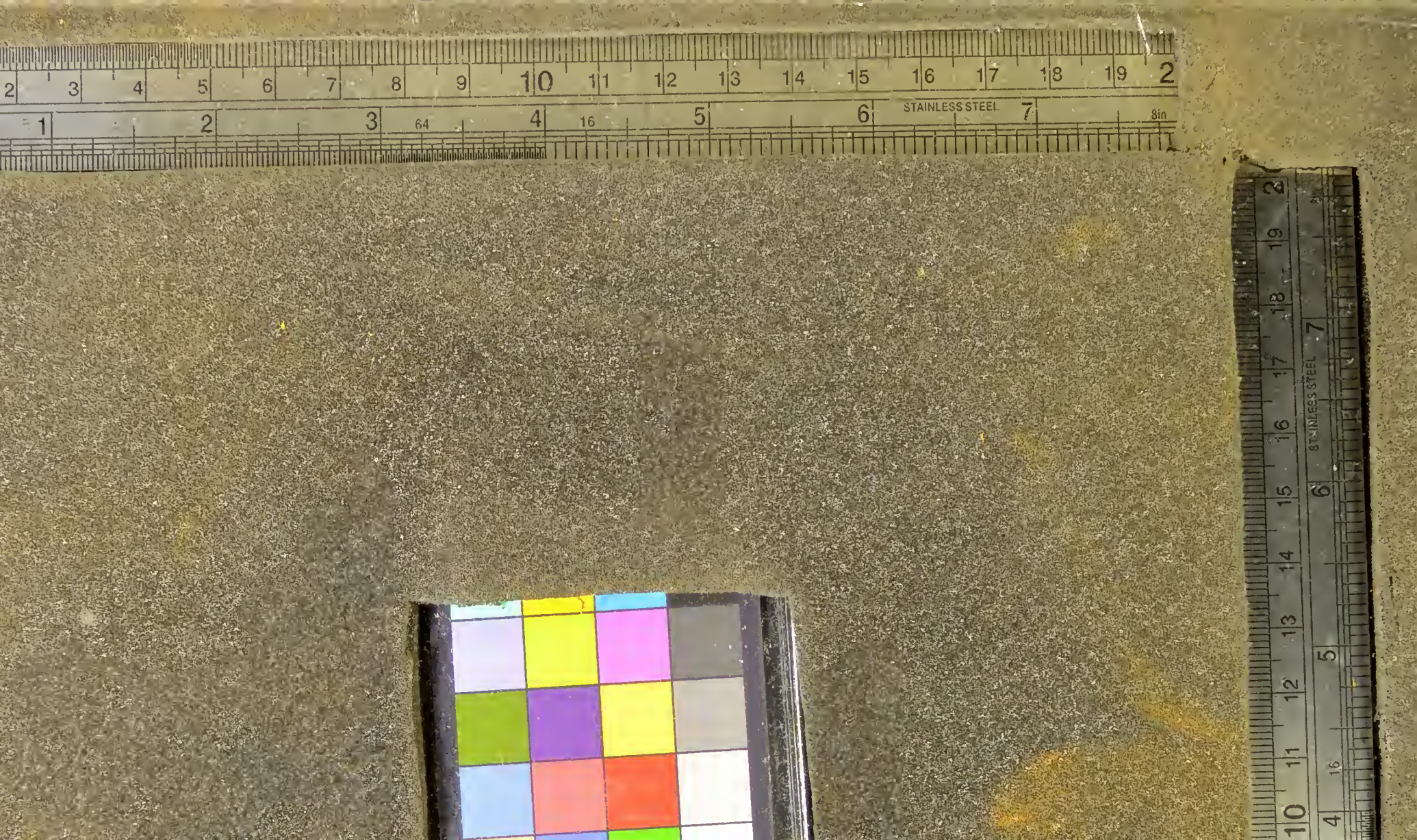
[Read before the Society, 12th May, 1886.]

The varieties of mal-development which have been described in connection with the genito-urinary organs are very numerous, and in many instances the products of abnormal action are so complex that explanation is difficult, often doubtful. The details of the case about to be described, while interesting in themselves and in respect of their apparent causation, are also noteworthy as presenting, when taken together, a marked approach towards the conditions which accompany the more complicated cases of vesical extroversion. It will be seen that the descriptions which follow are in many respects so similar that they suggest at least that the different cases had a similar origin.

The subject is a female fœtus of about the eighth month of intra-uterine development, kindly put in my possession by Dr. Grange of Cumnock. The length of the body from vertex to coccyx is $9\frac{1}{2}$ inches. The upper limbs and upper portion of the trunk are normal, but below the umbilicus many indications of mal-development are met with, the special details of which may be described under the headings of the tissues or systems which they affect.

Bones and Ligaments.

Both feet are clubbed—the right one particularly so. The right tibia shows an intra-uterine fracture, the end of the upper fragment forming a marked projection underneath the skin as may be seen from the plate. The right and left portions of the symphysis pubis are separated from one another to the extent of $1\frac{3}{4}$ inches, and, in consequence of this, the umbilicus is dragged downwards to a position much lower than usual, lying beneath a line drawn between the anterior superior iliac spines. In consequence, also, the lower limbs are widely everted. There is,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however, no rupture of the skin. A broad tumour over the sacrum owes its presence to a spina bifida which implicates the sacral vertebræ. An abnormal looseness of the sacro-iliac synchondroses gives rise to a slight backward dislocation of the ilia. The anterior surface of the sacrum is slightly convex forwards. The lumbar part of the column presents a curvature towards the right side.

The effect of these alterations in modifying the capacity of the pelvis is very considerable. In a normal fœtus with a body length of $10\frac{1}{2}$ inches, 1 inch more than the present specimen, the greatest transverse pelvic diameter at the brim was $1\frac{1}{8}$ inches, and the antero-posterior measurement from the promontory of the sacrum to the symphysis reached $1\frac{1}{2}$ inches. In the present case, the mesial antero-posterior diameter is reduced to $\frac{1}{4}$-inch; while the transverse measurement, which is less interfered with, marks under 1 inch.

Anterior Abdominal Wall.

The recti muscles arise from the widely-separated pubic bones, and are consequently very far apart from one another below. As they pass upwards, however, they also reach inwards, and after gaining the level of the umbilicus their internal margins are almost in contact. They are somewhat broader than usual. Strong bundles of fibres pass from each pubic spine to the umbilicus, forming two sides of a triangle, the base of which is represented by fibres of similar strength stretched between the portions of the cleft symphysis. The intervening space is filled up by fibres crossing between these bands, not so dense in character nor so regular in disposition. The middle portion of the anterior abdominal wall below the level of the umbilicus is thus closed in by fibres which extend between the widely divergent portions of the linea alba, and which in the normal state are not represented.

Intestine.

The duodenum takes an exceedingly sharp curve, the limbs of which lie in very close proximity. The upper descends from the pylorus and passes to the right side; the lower ascends towards the left to its termination, where, opposite the middle line, it is closely bound to the posterior abdominal wall. Owing to the sharpness of the curve, the pancreas is somewhat dislocated from its usual position, and shows as it crosses a marked convexity downwards. Its duct enters the duodenum separately almost half

an inch nearer to the pylorus than the hepatic duct. The jejunum immediately enters the mesentery, the line of attachment of which is directed downwards to the left iliac fossa instead of to the right. There is no part corresponding to the ascending tranverse and descending colon, but the ileum expands suddenly into a wide and irregularly-sacculated tube which passes downwards on the left side into the pelvis, the mesentery merging into a loose meso-rectum.

Examined closely, the tube into which the ileum passes presents four irregular pouches on its attached or mesenteric border. Beyond the last pouch the lumen of the tube becomes considerably reduced, and the intestine leaves the abdomen to enter the pelvis. The pouches possibly represent nothing more than an irregular dilatation of the least resistant part of the tube following upon the atresia of the anus, to be afterwards noticed. But possibly they may be due to the natural tendencies of growth of suppressed portions, the first representing the caput cæcum, and the others portions of the colon. In relation to this question, it is interesting to note the disposition of the blood-vessels. The ileo-colic artery supplies the lower end of the small intestine and the first diverticulum or pouch. The right colic supplies the second and a portion of the third. The remaining portion of the third and a part of the fourth are supplied by the middle colic artery. The inferior mesenteric completes the supply and furnishes branches also to the first part of the rectum. (The letters *b b* in the Plate represent the dilated portion of the intestine).

It is interesting to note that in spite of the absence of the transverse colon, the great omentum occupies its usual position. Its returning layers are fixed upon the left side to the abdominal wall along the usual line of the meso-colon, and in the middle line to the left surface of the mesentery.

One among a number of otherwise trivial vascular anomalies may be mentioned here—the right hypogastric artery is absent.

Genito-urinary Organs and Pelvic Viscera.

The kidneys are smaller than usual, and their surfaces are smooth and show no lobulation. The right occupies its usual position, but the left is displaced downwards to such an extent that its upper end lies on a level with the lower extremity of its neighbour. The inferior portion of this kidney passes into the

pelvis and is much disorganised evidently from the effects of pressure. The right ureter is, at its upper end, normal, arising from the hilus of the kidney and passing downwards on the inner side of the organ, but the left ureter springs from the dorsal surface of the left kidney and courses downwards upon its outer side. A portion of the left ureter also projects further up than the superior extremity of its kidney in the form of a small rounded sac. Both ureters end in a peculiar manner. They pass downwards underneath respectively the right and left horns of a bicornate uterus, and, turning round the outer margins of the horns, become incorporated with their walls on the anterior surface. In the course downwards from the kidney to the uterus the lumen of each is gradually lost so that the tubes are reduced to solid cords before they sink into the uterine substance. The cords may be traced downwards for some distance along the front of the uteri by dissection into the walls. They will be again alluded to in connection with the vaginæ. (The letter *c* in the Plate is placed upon the right kidney, and the ureter stretches between *c* and *d* to end upon the uterine horn.)

Both ovaries are present. Each is about half-an-inch in length. and is rather narrow. It is attached by a fold of peritoneum to the Fallopian tube which lies immediately above it (*e* in the Plate). A firm round ligament connects the ovary with the uterine horn.

Each uterine horn *(d. d.)* is a tubular structure, containing a long narrow cavity. It is distinguished from the Fallopian tube above, and the vagina below, by its greater thickness. The Fallopian tube, uterus, and vagina upon each side, thus form a continuous tube-like structure, the middle portion of which, the uterus, is thickest, and contains a cavity, the upper portion, or Fallopian tube, thinnest and impervious, while the vagina, intermediate in thickness and somewhat flattened from side to side, is separated from the uterus by a marked construction, and is also impervious. A well-marked round ligament upon each side passes from the upper end of the uterine horn through the abdominal wall in the usual way.

The bladder, the lower ends of the two vaginæ, and the lower end of the rectum, are firmly bound together, and form a compact mass situated in the pelvic cavity. They apparently open externally by a single aperture, the position of which is rather further forwards than that which the vulva occupies in normal circumstances. The opening is less than $\frac{1}{4}$ inch in length, and is

surrounded in front and on both sides by prominent folds of skin, which behind merge into the flat margins of a shallow groove, continued backwards between the thighs, to be lost upon the swelling occasioned by the sacral spina bifida.

The anterior wall of the bladder is firmly incorporated with the tissue which has been described as filling up the gap upon the anterior abdominal wall, between the divided portions of the linea alba. The upper portion of the posterior wall is free, but into the lower portion the two vaginæ, which now join with one another, seem to pass directly. To the back of the vaginæ, the rectum is firmly fixed.

When the bladder is laid open by the mesial division of its anterior wall, it is found to contain a cavity which opens widely below. This opening is the entire cloacal aperture before alluded to, which is therefore not subdivided into genital and urinary portions, but passes directly into the bladder. The anterior surface, the fundus, and the upper portion of the posterior surface of the cavity are lined with a whitish membrane, evidently the vesical mucous membrane. This membrane is deficient over the lower portion of the posterior wall, but in front and at the sides it sweeps down to the margin of the cloaca. There is thus left an area in the shape of a horse-shoe upon the base of the bladder, which is left uncovered by the membrane which lines the rest of the interior. Upon examining the uncovered area, it is noticed that it is separated into two portions, an upper and a lower, by an aperture placed between (above and to the left of *f* in the Plate). This aperture is a fistulous communication with the rectum. The portion above it is brownish in colour and soft, the portion below is of a whiter colour and of a firm membranous character, and forms the posterior wall of the cloacal outlet. The impervious vaginæ meet as they reach the back of the bladder. Their more anterior fibres end upon the brown surface above the rectal fistula, while the most posterior fibres descend by the margin of the opening, to form by their junction in the middle line below it the firm whitish area which completes the cloaca behind. The ureters may be traced down upon the anterior vaginal walls for some little distance, but cannot be followed quite to the bladder; their fibres, doubtless, reach the base along with the anterior vaginal fibres just described. The rectum passes down behind the vaginæ where they unite with one another at their lower ends, forming a firm adhesion to their posterior surface, and by means of

a perforation extending through the vaginal tissue its cavity is put into communication with the bladder. Still further down the rectum forms an impervious cord which reaches the skin behind the cloaca.

The cloacal outlet represents, therefore, a genito-urinary aperture only, the position of the impervious anus behind it being marked by the blind termination of the gut. The anterior wall of the opening is urinary, being continuous with the anterior wall of the bladder; while the posterior wall is genital, continuous with the vaginal fibres. There is no subdivision into urinary and genital portions. At the same time, the base of the bladder and the posterior urethral wall are wanting, and the anterior vaginal fibres are not prolonged down to the margins of the outlet.

Causation.

Many of the abnormalities present, both in themselves and in their relations to one another, are easily explained. The talipes, intra-uterine fracture, spina bifida, lumbar curvature, looseness of the synchondrosis, with the consequent displacement backwards of the ilia, and separation of the component parts of the symphysis, are all evidences of an interference with the normal development of the bony and ligamentous tissues of the lower part of the body. Inflammations even of a localised and insignificant nature are known, when they occur in early embryonic life, to produce results by their interference with developing structures quite out of keeping with their apparent character. It is exceedingly possible that the same exalted or inflammatory action at a very early period of intra-uterine life, which gave rise to the spina bifida, affecting the sub-cutaneous or sub-peritoneal tissues and setting up irritation, instituted changes in the lower portion of the body, with the effect of disturbing the relations of the bony and ligamentous parts.*

The condition of the anterior abdominal wall—the linea alba being split into two, the gap bridged by fibrous tissues—seems to be referable to rupture, slowly produced, consequent on the separation of the pubic bones. The position of the bladder as it reaches to the umbilicus is one of close apposition to the abdominal wall,

* The manner in which very slight stimulations or irritations may affect the whole development of the body is dwelt on by Professor Cleland, "Contribution to the Study of Spina Bifida," &c. *Journal of Anatomy*, vol. xvii., p. 257.

and in the case of rupture the anterior vesical wall would naturally fill the gap, forming adhesions with the torn edges. In this way the incorporation of the wall of the bladder with the fibrous tissue which crosses the gap receives a simple explanation. It is possible to imagine that, after adhesion in this manner, the causes to which the original rupture was due might still operate with the effect of destroying the anterior wall of the bladder also, and causing its posterior surface to appear upon the abdominal parieties.

The intestines and genito-urinary organs bear traces rather of the arrest of the normal progress of development than of positive abnormal action, and this arrest is in all probability due to the interference with the growth of the containing cavity and to pressure. The arrangement of the intestine, the bicornate uterus, the double vaginæ, the small size of the kidneys, and the impervious ureters, are all examples of such a condition. The atresia of the anus is probably due to the pelvic deformity, and to the encroachment of the tumour of the spina bifida. The incorporation of the ureters with the uterine walls is interesting. It is possibly brought about here simply by pressure; but in another case of a fœtus affected with genito-urinary abnormality (in which, however, there was no suspicion of undue pressure) I noticed a connection somewhat similar in character, though of less extent, upon the right side. Boogaard* has described and figured a case in the male in which a persistent Müller's canal was in like manner connected with the coats of a ureter, although their respective cavities did not enter into communication.

The absence of the trigone of the bladder and the posterior wall of the urethra, and the non-development downwards to the outlet of the anterior vaginal fibres, giving to the posterior true vesical wall an appearance of being bifid below, and setting the cavity into wide communication with the cloaca, are possibly also evidences of arrested development. But if this be the case, it is impossible to apply our present notions of the development of these parts to the details of its explanation. Nor, upon the other hand, is it likely that simple fistula could have produced such a complete fusion of the genital and urinary cavities in their lower parts as is here present. The difficulty seems to me to arise from an erroneous conception of the mode of development of the lower part of the bladder—a subject which, however, I shall leave

*Verslagen in mededeelingen der Kon. Akademie van Wetensch-Afdeel. Natuur Kunde, 2e reeks, 9e deel. Also, *Journal d' L'Anatomie* 13.

for consideration until the deformities of a similar nature described in allied cases have been briefly noticed.

Vesical extroversion.

The peculiar feature in this interesting abnormal condition is the appearance of the posterior wall of the bladder upon the front of the abdomen, this being due to a deficiency alike of the anterior abdominal and vesical walls; but in the great majority of cases this malformation does not stand alone, but is accompanied by others of a more or less important character.

The literature of extroversion is exceedingly large. A succinct account of the main details of the deformity is given by Phillips.* And among the many other more recent contributions is one by Champneys† in which the author, after describing a special dissection, reviews very thoroughly the whole subject, and gives an elaborate list of the writers who have already treated it. Still more modern additions to the literature will be found in the description of special cases by Doran‡ and Ogston.§ The general summary which will be given here is in great part drawn from Champneys' paper, and the references to the descriptions cited will be found in the very complete list of works with which that paper is furnished.

The varieties of the characteristic malformation are very numerous. Mayo describes a case in which there was no fissure either of the abdominal or vesical wall, but only a hernial pouch containing a perfect bladder. The pubic bones were five inches apart. Vrolik describes a case in which a perfect bladder protruded through a fissure of the anterior abdominal wall. On the other hand, the posterior wall of the bladder is not only found projecting upon the surface of body, but cases are described in which the vesical surface is partially or completely split into two lateral portions, the intestine or genital organs lying between the parts. Of such a nature are those described by Bartels, Retzius, Friedlander, Rose, Fränckel, Meckel, and Doran. So far as I have been able to determine, in cases of complete fission it is usually only the lower portion of the normal posterior wall which

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* Todd's *Cyclopæd. of Anatomy and Physiology*. Vol. i. Article Bladder.

† St. Bartholomew's Hospital *Reports, 1877.*

‡ *Journal of Anatomy and Physiology, 1881.*

§ *Journal of Anatomy and Physiology, 1882.*

is present as the divided body, the urinary and generative openings being placed towards the upper extremity of each half. The ureters, when they are not impervious, usually open upon the extroverted surface, in the simpler cases towards the lower end. The vagina, which is very often double, frequently opens immediately beneath it, or between the portions, and even in some very complicated cases upon the vesical surface. The vagina, however, is often quite normal in position.

Champneys' case is very interesting in respect of these openings. The ureters opened separately towards the lower end of the mass, their openings being overhung by prominent wrinkles, suggesting the valvular folds of normal circumstances. Two vaginal openings were placed immediately beneath, overhung in their turn by the projecting lower border of that part of the mass upon which the ureters opened.

The pelvic and abdominal viscera are usually interfered with. The anus often terminates blindly, or there may be fistulous openings into the vagina or upon the anterior abdominal wall, between the lateral parts of the cleft tumour. The ureters are often dilated, sometimes impervious. The kidneys in like manner are frequently affected. The usual arrangement of the intestine is in most cases interfered with, and its peritoneal relations changed. One hypogastric artery is usually atrophied. The penis is frequently split.

There are in almost every case evidences of defects in the bony and ligamentous tissues, particularly those of the pelvis. The symphysis pubis is usually cleft. At one time this was held to be invariable, but sufficient proof has been adduced to show that it is not absolutely constant. Dislocation backwards of the ilia is frequent. Spina bifida is very common; Champneys' and Doran's cases both show examples. It usually affects the sacral or lumbar vertebræ. Curvature of the spinal column is one of the most frequent co-existing deformities. Evidences of similar defects in the bones and ligamentous tissues are usually present in other parts of the body also. Talipes is very frequent, and in connection with the bones of the skull hare-lip and non-closure of the vault have been described.

The theories which have been applied to the explanation of this deformity are exceedingly numerous. Champneys, in his interesting paper, quotes 17 different views, and adds himself one more. It is needless to go into details respecting the many opinions of

the different writers upon the subject. The most interesting are those of Duncan, Velpeau, and Isidore G. St. Hillaire. Duncan regards the deformity as due, in the first place, to an imperforate condition of the urethra. Distention of the bladder follows on account of the accumulation of urine, divarication of the pubic bones and rupture of the abdominal and vesical walls follow in sequence, and thus extroversion is produced. It is hardly necessary to refute this theory. The urethra is not found impervious as a rule, nor does the fœtus secrete sufficient urine to distend the bladder to such an extent; and if it did, distension of the bladder is not likely to cause rupture of the abdominal walls.

Velpeau believes it to be due to a process of ulceration from disease, implicating the parieties of the abdomen and bladder. Such a theory does not account for the allied deformities which are so typical of the abnormality. He maintains that the pubic bones are not simply separated but destroyed; but this has not been found to be the case.

Isidore St. Hillaire simply refers it to an arrest of development, and does not attempt to specify the exact manner in which the special anatomical peculiarities are produced. He bases his arguments upon the unmistakable instances of arrested development which invariably accompany the malformation. There is doubtless a general truth in St. Hillaire's theory, but it lacks in detail. Champneys accepts in a general way a theory of arrested development, but seeks to work it out into details, more especially to make it explain the bilateral fission of the extroverted mass which has been noticed as occurring in many cases. According to him the lower abdominal walls have failed to unite from arrest of development. The cleft bladder he explains by regarding the allantois as taking its origin from two lateral portions which subsequently unite to form one vesicle. In this view Champneys follows Bischoff,* who thus assigns a double origin to the urinary bladder.

The explanations offered by Champneys cannot, however, be accepted as satisfactory. The arrest of development is assumed, and no theory of its causation is attempted, nor is anything suggested to explain the very special manner in which the lower abdominal and the vesical walls suffer. In addition, embryologists are now agreed in regarding the allantois as originally a single

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* Entwickelungs-gesch. d. Saugethiere u. d. Menschen, Leipzig, 1842.

structure, and Bischoff's views have not found confirmation in modern research.

Reviewing the details of the undoubted cases of extroversion, and comparing them with those of the dissection which I have described, it will be noticed that the latter approach the former in many respects very closely. The accompanying malformations in my case are those which are found associated with the most exaggerated forms of extrophy. Further, although the abdominal and vesical walls are not ruptured, they yet present appearances from which it is reasonable to infer that the causes which have operated in producing their abnormal condition would, if prolonged or heightened in their action, have resulted in complete cleavage. Finally, had destruction of the anterior wall of the bladder taken place, the posterior wall would have shown, upon the front of the abdomen, a surface bifid below, the rectum opening by a fistula through the vaginal fibres between the lateral portions. I am inclined, therefore, to assign to extroversion a causation similar to that already detailed—interference with the bony and ligamentous portions of the pelvic walls due possibly to early fœtal movements, following upon irritation or inflammatory action, and consequent alterations in the shape and dimensions of the cavity.

The mode in which the rupture of the abdominal wall and the destruction of the anterior wall of the bladder take place, has already been sufficiently indicated, and the effect of the interference with the growth of the pelvis in arresting the development of the organs contained in it has been pointed out. Cases are doubtless described in which extroversion was present unaccompanied by cleavage of the symphysis, but it is possible to imagine causes which would produce great lateral stretching, even with intact articulation.

There remains for consideration still, the method by which the apparent fission of the extroverted mass into two portions is produced. From my dissection it is evident that the bifid appearance of the posterior vesical wall is due to the absence of the trigone and the urethral floor, and in extroversion when fission is present the constant position of the genital or anal apertures between the portions, points to a similar condition. The accepted view,* which ascribes to the whole of the bladder and to the first portion of the urethra, an allantoic origin is

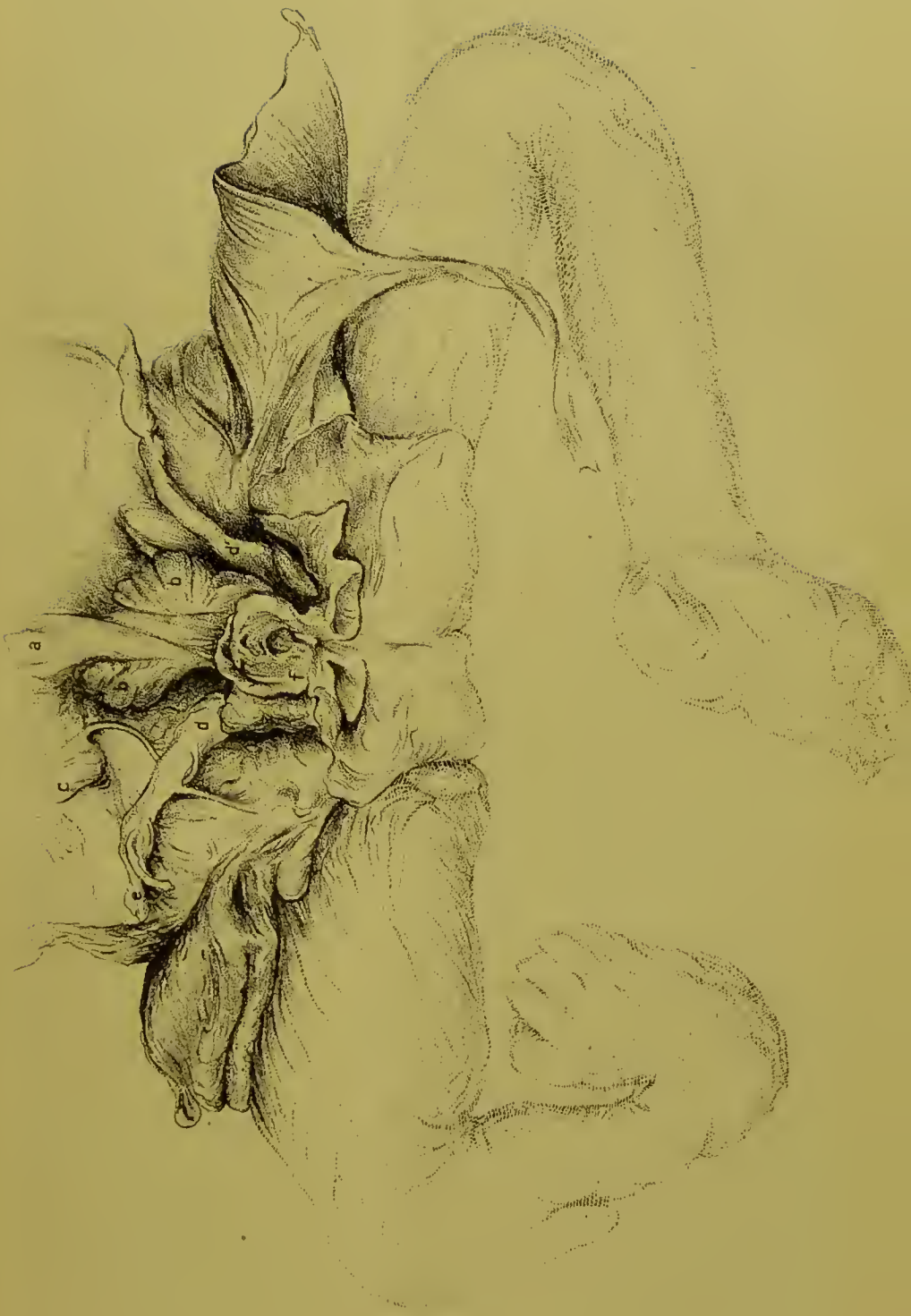
* See *Comparative Embryology.* F. M. Balfour, London, 1881.

unsatisfactory, inasmuch as it does not explain the method by which the ureters are transferred from their original termination on the cloacal end of the gut to the back of the bladder; while the manner in which the urinary and genital ducts are separated from one another at their lower ends is still regarded as an open question.

I am inclined to suggest a hypothesis of the development of the lower end of the bladder somewhat different from the usual one. The cloacal end of the gut is, in a manner not yet accurately known, subdivided into an anterior or genito-urinary portion, and a posterior or intestinal portion, which open separately upon the surface. Into the genito-urinary sinus the allantois ureters and genital ducts open in order from before backwards. Apparently what happens is, that the *tissue between the ureters in front and the genital ducts behind is prolonged down, so as to form a septum, the anterior surface of which persists as the trigone of the bladder and the floor of the first portion of the urethra, while its posterior surface is vaginal.*

Apart from embryological evidence, many things render this probable. Among these are the constant position of the ureters at the upper end of the trigone, the absence of peritoneal cavity between the trigone and the vagina or rectum, and the exceedingly close anatomical connection which subsists between the walls of the lower parts of these organs. Many cases in abnormal anatomy are described, in which ureters or genital ducts opened into cavities other than the usual; and, while some of these are possibly to be explained by fistula, there are others to which that explanation cannot be applied. These can only be understood on the supposition that the septa have developed in an irregular manner.

In my dissection, and in those cases of extroversion in which partial fission is found, the suppression of the septum between the genital and urinary portions, and the consequent absence of the base of the bladder and anterior vaginal wall, explain the bifidity of the lower portion of the vesical wall, and refer it to arrest of development, other instances of which are numerous. Complete fission of an extroverted tumour may be explained on the supposition that the upper portion of the posterior wall of the bladder, as well as the anterior wall, has suffered destruction, the protruded mass representing the lower lateral portions, which, in normal circumstances, bound the sides of the trigone.



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DESCRIPTION OF PLATE VII.

a. Small intestine.
b. Diverticula from large intestine.
c. Right kidney.
d. d'. Right and left uterine horns.
Between *c* and *d* the right ureter.
e. Right Fallopian tube.
f. Posterior aspect of bladder.

Above and to the left of *f* a round aperture indicates a fistulous communication with the rectum.

Above the opening, the limits of the vesical mucous membrane are marked by a horse-shoe shaped edge.